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CASE REPORT

Cranial fasciitis in an infant

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Summary Cranial fasciitis is a rare reactive, non-tumoral lesion (pseudosarcoma) that occurs mostly in children below 6 years, with a propensity for the cranium. This type of tumor has been reported to arise from the deep fascia, periosteum or from the fibrous layers that cover fontanelles and sutures. The lesion is usually solitary, firm and painless. Grossly, it is usually unencapsulated but well circumscribed. To our knowledge, only 51 cases have been reported in the literature. Herein, we present the clinical, pathological, and radiological findings of a female infant with cranial fasciitis of the skull, and review the literature.

1. Introduction

Fibroblastic and myofibroblastic lesions are inflammatory reactions including proliferative fasciitis, proliferative myositis, nodular fasciitis, intravascular fasciitis and cranial fasciitis of childhood. They were first described by Konwaler and Weiss in 1955 and the lesions in most cases occurred after trauma.[1,2] Such lesions are usually misdiagnosed as sarcomas because of their clinical and histological appearance.[2,3]

Cranial fasciitis is considered a subset of deep nodular fasciitis and is a non-neoplastic benign subcutaneous condition of the head.[4] Cranial fasciitis was first described by Lauer and Enzinger in 1980 based on the pathological findings in nine cases with ages ranging from 3 weeks to 6 years.[1,5] It is usually unencapsulated but well circumscribed. Clinically, cranial fasciitis usually grows rapidly up to a diameter of 1–3 cm or more, representing a solitary, firm and painless skull mass.[5] The etiology of this lesion is still unknown. It is thought to be a reactive proliferative process, although antecedent local trauma is usually lacking.[2] The average age of onset is 2 years. The male/female ratio is 2:1.[6]

To the best of our knowledge, 51 cases have been reported in the literature.[7,8] We hereby add another case of cranial fasciitis in a 7-month-old female with the clinical course, histological appearances and the radiological findings.

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2. Case report

A 7-month-old girl presented with a palpable, round mass in the right frontoparietal region. According to the statement given by her family, the patient did not seem to have any discomfort and the mass had been present for about 1 month. The patient was born at 39 weeks' gestation via spontaneous vaginal delivery without any assistance, and her family denied any history of trauma.

Physical examination revealed a nontender, well-circumscribed mass in the right frontoparietal region measuring about 2 × 2 cm. The lesion was firm and there was no skin lesion over the mass. There was no neurological deficit.

A skull X-ray (Fig. 1A) revealed an approximately 1 cm ovoid osteolytic lesion with a thin sclerotic border in the right frontoparietal region. An unenhanced computed tomography (CT) scan of the brain (Fig. 1B) showed an approximately 1 cm osteolytic lesion involving both the inner and outer tables of bone with a soft tissue component in the right frontoparietal bone without intracranial invasion. Magnetic resonance images or MRIs (Figs. 1C and 1D) demonstrated a bone lesion of intermediate signal intensity in the right frontoparietal region on T1-, T2- and diffusion-weighted images. This feature was thought to be compatible with a predominantly fibrous lesion without abnormal diffusion restriction. The initially radiological diagnoses included Langerhans cell histiocytosis, epidermoid and dermoid tumor.

After being transferred from the pediatric department, the patient underwent gross-total resection (Fig. 2B) because of the osteolytic appearance of the lesion in the brain CT scan. Intraoperatively, the mass was found to extrude through the center of the exposed bone (Fig. 2A). The lesion was soft and fleshy, and easily dissected off the dura with blunt dissection. Histopathologically, the tumor was composed of proliferated spindled to ovoid cells with low to moderate cellularity. The cells were arranged in sweeping fascicles with focal myxoid feathery background. Vascular proliferation, erythrocyte extravasation and collagen matrix precipitation were present. Nuclear atypia was subtle, mitoses were rare, and necrosis was absent. In

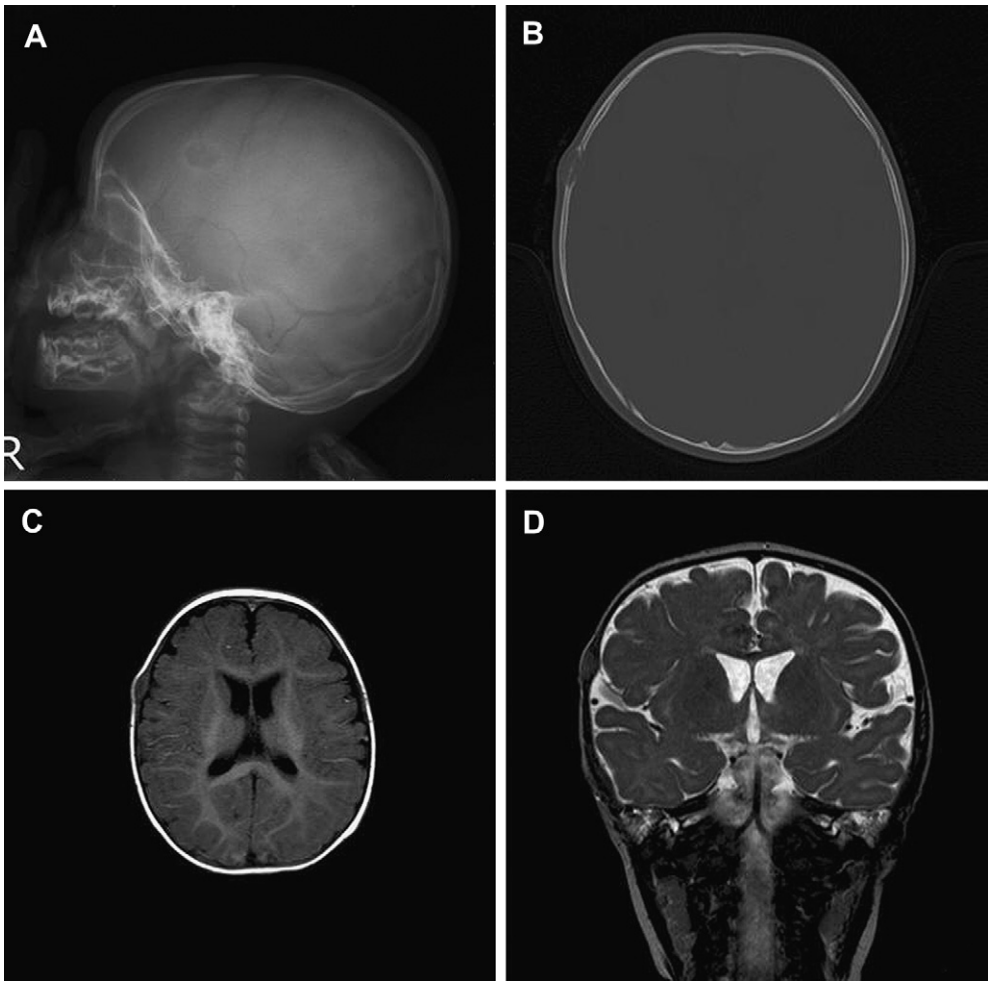

Figure 1 Radiological pictures. (A) Skull X-ray showing a 1-cm ovoid osteolytic lesion with thin sclerotic border in the right frontoparietal region. (B) An unenhanced CT scan of a brain showing a 1 cm osteolytic lesion involving both inner and outer tables with soft tissue component in the right frontoparietal region without intracranial invasion. (C) and (D) Magnetic resonance images demonstrating a lesion with intermediate signal intensity in the right frontoparietal region on T1-, T2- and diffusion-weighted images.

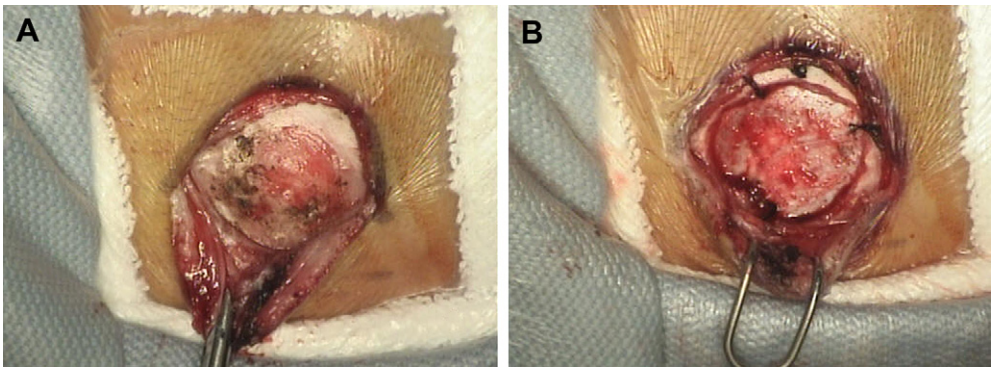

Figure 2 Intraoperative pictures. (A) Mass found to extrude through the center of the exposed bone. (B) After gross-total resection.

an immunohistochemical study, the lesional cells were positive for smooth muscle actin (SMA), but negative for desmin, S-100 protein, CD34 and epithelial membrane antigen (EMA) (Fig. 3). Based on the results of immunostaining and the histopathological features, a cranial fasciitis, variant of nodular fasciitis, was diagnosed.

3. Discussion

Cranial fasciitis is a very rare disease first described by Lauer and Enzinger in 1980.[5] The lesion comprises only 1% of all pediatric subscalp tumors.[1] A review of the literature shows only 51 cases to date,[1] and Table 1 summarizes 52 cases of cranial fasciitis, including our patient.

Cranial fasciitis is a fibroproliferative lesion of the scalp most commonly seen in the very young pediatric population. The histological findings of cranial fasciitis are similar to those of nodular fasciitis. The differences between these two lesions are the location and age predilection. In 1955, Konwaler and colleagues first described nodular fasciitis as a benign infiltrative disease of the subcutaneous tissue, muscle or fascia occurring anywhere in the body.[9] Nodular fasciitis is the most common pseudosarcomatous lesion of the soft tissue. It is also called proliferative fasciitis and is most commonly seen in patients between the ages of 20 and 35.[5,10] Nodular fasciitis has been referred to as apseudosarcomatous lesion in appearance, due to the presence of histological findings such as high cellularity and occasional mitotic figures.

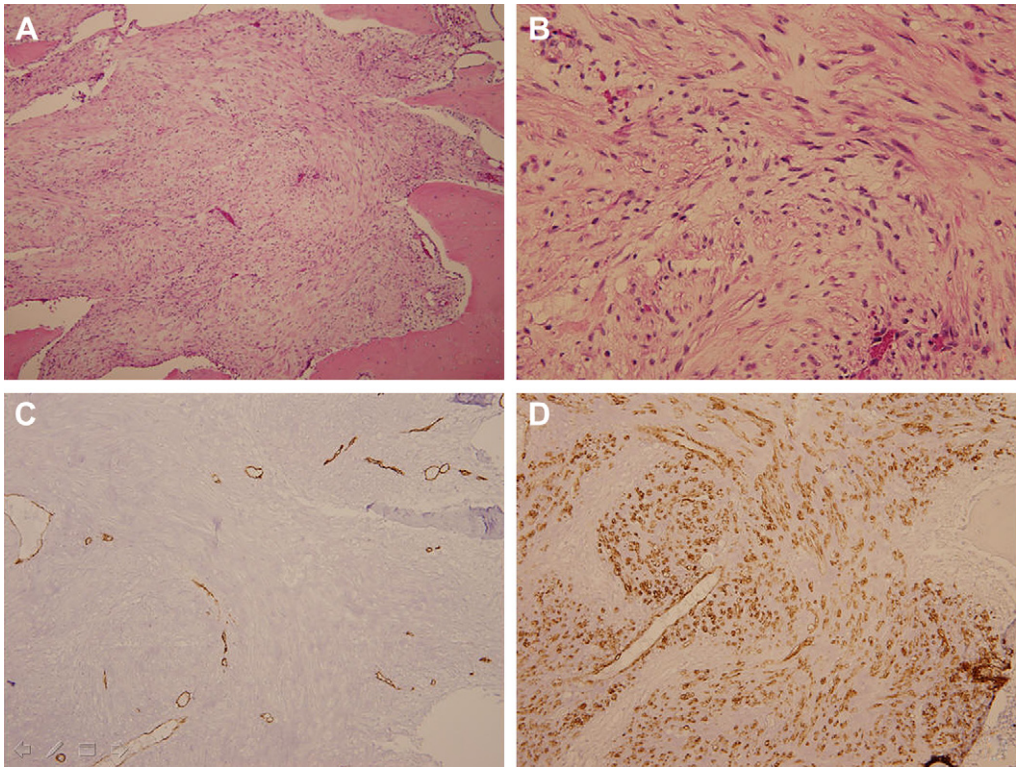

Figure 3 Histological findings. (A) H&E stain 100×. (B) H&E stain 400×. (C) Immunohistochemical study negative for CD 34. (D) Immunohistochemical study positive for smooth muscle actin (SMA).

Table 1 Summary of 52 cases with cranial fasciitis.

Male:female	34:18 (2:1)
Age	From day 1 to 47 y
Average	34.3 mo, excluding 3 adults
Males	26.1 mo
Females	50.1 mo
Location	
Temporal	17
Frontal	13
Parietal	6
Other location	12
Intradural extension	3 (6%)
History of head trauma	
Yes	10 (19%)
No	19 (37%)
Unmentioned	23

Cranial fasciitis is a subset of nodular fasciitis.[11] Both conditions consist of predominantly spindle-shaped and stellate fibroblasts with some mitotic figures. Due to the clinical course and histological findings, these lesions are potentially capable of evoking the erroneous diagnosis of sarcomas. An improved understanding of clinicopathological features and the biology of cranial fasciitis is therefore required to help us prevent unwarranted wide surgical resection and adjuvant therapies.

Cranial fasciitis normally presents as a firm, nontender, well-circumscribed lesion. The most common site of the lesion is temporoparietal area,[12] but it has also been reported in other locations such as the anterior fontanelle, cheek, forehead, orbit, maxilla, petrous bone, posterior auricular region, occiput, vertex, and the ear.[11] The lesions typically grow rapidly and are rarely larger than 5 cm in diameter, but may attain an impressively large size.[13] The exact origin is not clear, but most authors believe that the lesion arises from the deep fascia, periosteum or the fibromembranous layer covering sutures and fontanelles, and often erodes only the outer layer of the skull, typically invading the overlying scalp.[7] Invasion of both layers of the skull with dural invasion and compression of the brain has been reported.[14] A large epidural mass and exclusive intracranial extension with mass effect have also been reported, and even lesions causing significant compression of the superior sagittal sinus and underlying cortex have been described.[6,15]

The most common differential diagnosis is fibrous dysplasia, and distinction between the two conditions is often difficult.[11] Rapid growth and areas of dense cellularity with occasional atypical nuclei may be confused with sarcoma, but in cases of cranial fasciitis the lesions show no pleomorphism on microscopic examination.[11] The other differential diagnoses include sarcoidosis, infection, surface metastases, traumatic lesions, cavernous hemangiomas, myositis ossificans, Ewing sarcoma, fibrous histiocytoma, myxoid liposarcoma, myxofibrosarcoma, neurofibroma, meningioma, primary bone tumors, juvenile fibrosarcoma, and inflammatory lesions.[11,12]

Although there are no definite predisposing factors, up to 15% of patients have a history of local trauma.[11] There have also been reports maintaining that these lesions are related to birth trauma and prior craniotomy.[1,11,13] According to these reports, local injury or inflammation may trigger fibroblastic proliferation.[11] Rakheja and coworkers have mentioned that a dysregulation of the Wnt/β-catenin pathway possibly indicates a subset of cranial fasciitis, and this subset is pathobiologically related to desmoid fibromatoses rather than to nodular fasciitis. They also think that occasional cases of cranial fasciitis may be associated with familial adenomatous polyposis, which probably serves as an early indicator of this disease.[4]

Cranial fasciitis usually appears as a lytic skull lesion with or without sclerosis in imaging.[11,12] The lesion is well defined with an accompanying enhancing soft tissue mass seen on CT scans or MRIs.[16] CT scans may show an ossifying soft tissue mass with or without bone erosion.[17] MRI of the lesion usually reveals low to intermediate signal intensity on T1-weighted MRI, usually with enhancement, and intermediate to high signal intensity on T2-weighted images.[11,16]

Cranial fasciitis is a well-circumscribed, nonencapsulated tumor with spindle cells in a nodular pattern with proliferation of fibroblasts and myofibroblasts.[11] The histological characteristics of cranial fasciitis consist of dense areas of proliferated spindle or stellate-shape fibroblasts in a storiform or nodular pattern, mixed with loose areas representing myxomatous change.[11,14] Inflammatory cell infiltration and endothelial proliferation are occasionally seen. Mitotic figures and hemorrhage may be seen in some cases.[11] The immunostains were characteristically negative for S100 and glial fibrillary acidic protein. The lesion gives positive results in the tests for muscin, vimentin and muscle-specific actin, and in staining for collagen and reticulin.[11,18,19]

The choice of treatment for cranial fasciitis is surgical resection. Excision of the mass with local resection or curettage of the affected underlying bone is suitable in most cases and the recurrence rate has been reported to be around 1%.[11,20] Spontaneous involution has, however, been reported following fine needle aspiration, excisional biopsy or incomplete excisions.[5,21] Intralesional corticosteroid injection in three cases of cranial fasciitis presenting as subcutaneous nodules has been reported to bring about rapid and complete resolution.[20] Hass recommended that a "watch and wait" approach might be appropriate, especially for lesions in certain locations, such as the central face.[22] Nevertheless, the lesions usually grow progressively, and the rapid and destructive growth of these lesions can produce significant bony destruction and infiltration of adjacent structures with significant mass effect. They also have a favorable prognosis after resection, irrespective of whether it is complete or incomplete. Early excision might therefore be better than a "watch and wait" approach for large, rapidly growing masses.[23]

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